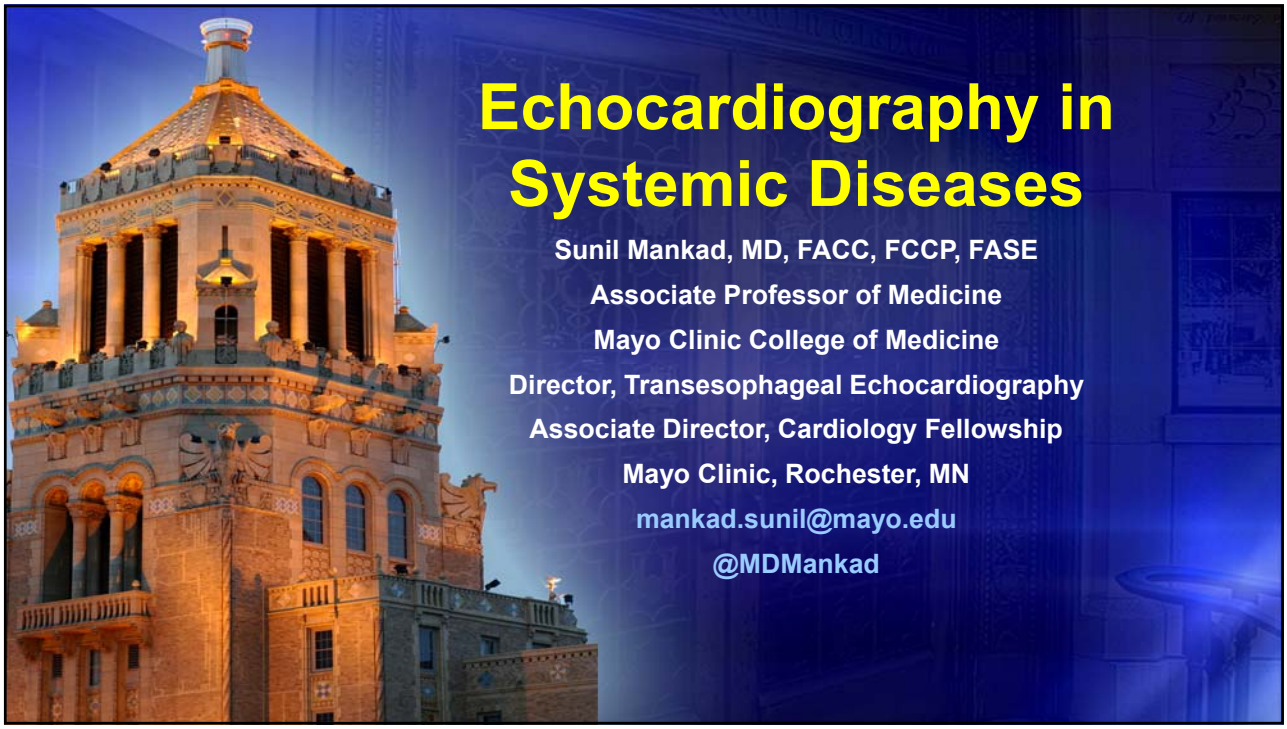

Echocardiography in Systemic Diseases

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DISCLOSURE

Relevant Financial Relationship(s)

Advisory Council: Siemens Healthcare, Ultrasound

Off Label Usage

None

Echo in Systemic Diseases

- Systemic diseases with secondary cardiac involvement are uncommon

But

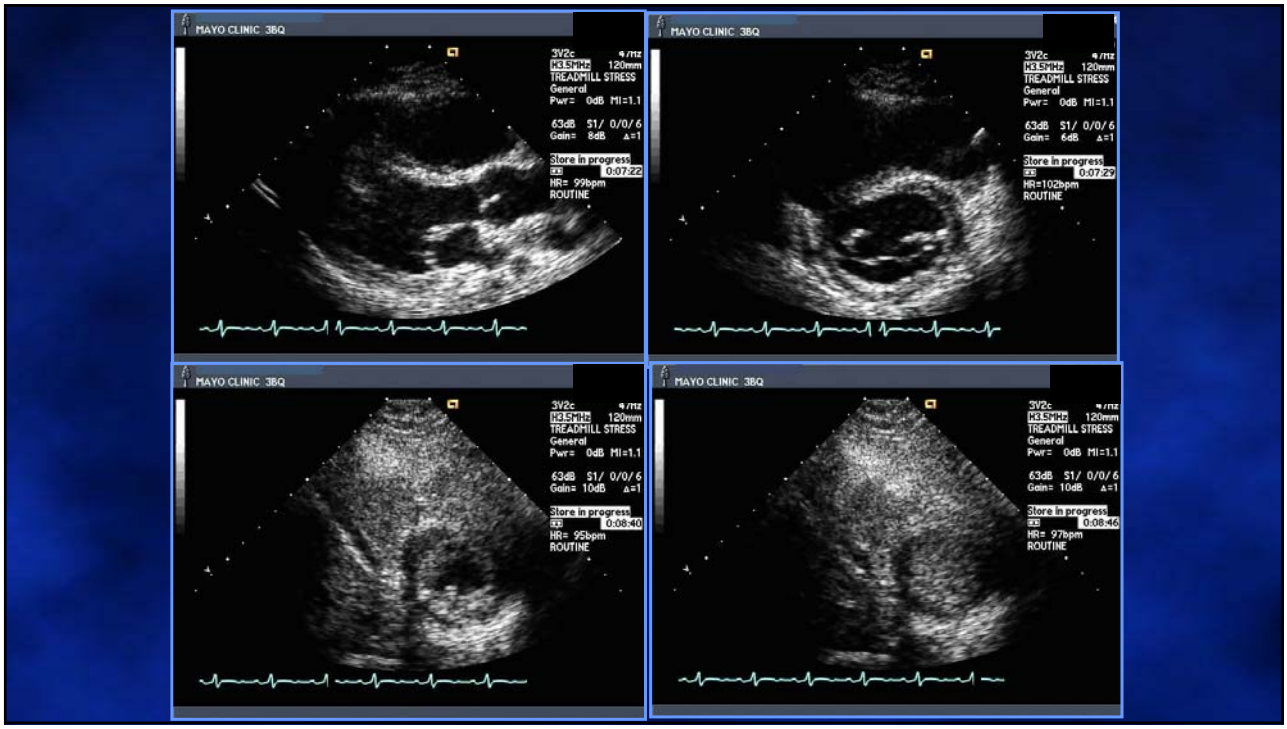
- Echo can identify unique, characteristic features and echo may be the first clue to the underlying systemic illness

Cardiac Involvement in Systemic Diseases

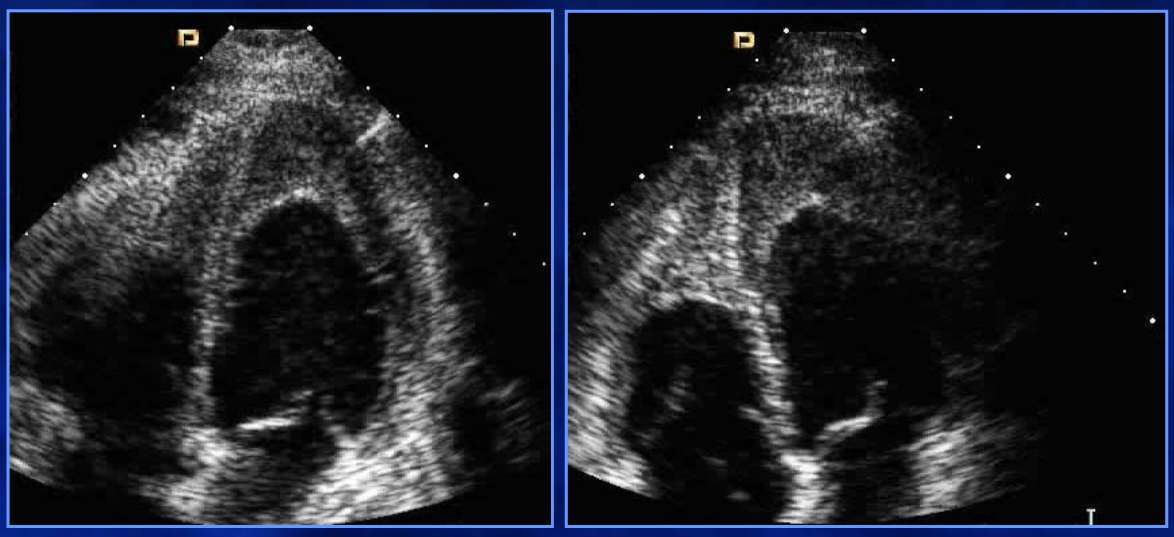
- Autoimmune
- Endocrine
- Collagen Vascular Diseases
- Malignancy
- Amyloid/Infiltrative Diseases
- Radiation Induced Heart Disease
- Drug Induced Valvulopathy

Case

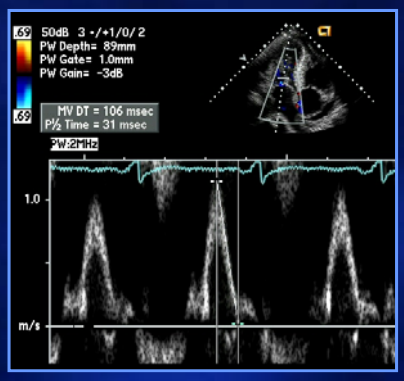
- 27 y/o female who presents with dyspnea, chest pain, and fatigue
  - NYHA class III
- Abnormal nuclear perfusion stress test led to coronary arteriography
  - Normal coronaries but LV gram suggestive of "Hypertrophic CM" (EF 75%)
- Elevated Sedimentation Rate
- Referred to Mayo Clinic → Echo performed

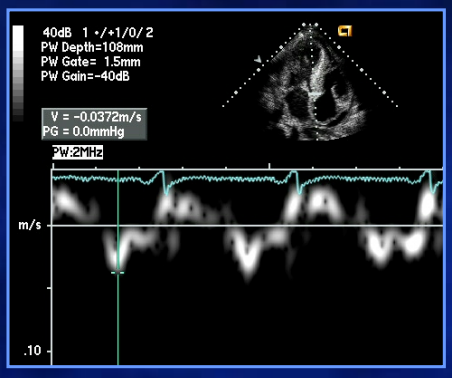


Apical 4 Chamber Views



Diastolic Function



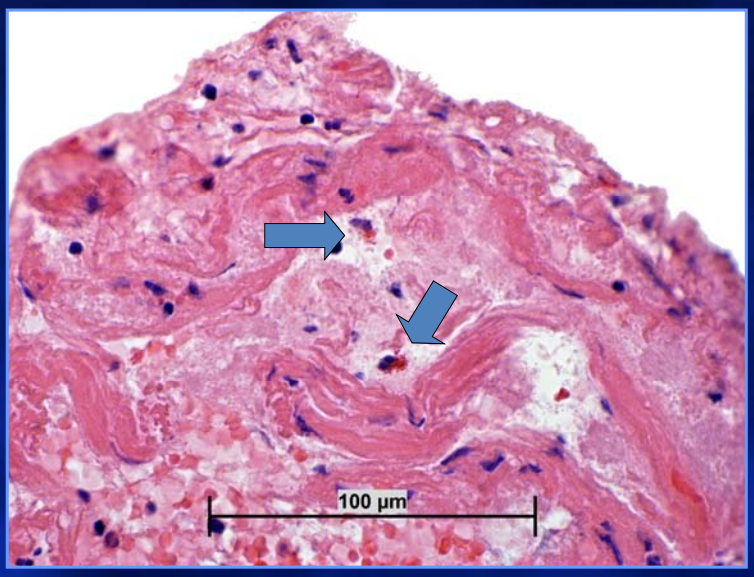


- MV Dec. Time = 105 msec
- MV Emax = 1.1 m/sec
- e' = 0.04 m/sec
- E/e' 28

What is the Diagnosis?

1. Hypertrophic Cardiomyopathy (Apical Variant)
2. Amyloidosis
3. Eosinophilic Endomyocardial Disease
4. LV Noncompaction
5. LV Myxoma

RV Biopsy (H&E Stain)

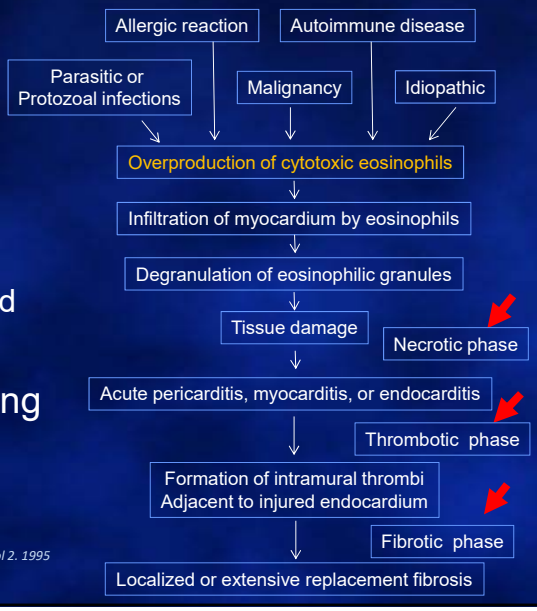


Hypereosinophilic Syndrome
Cardiac Manifestations

- Persistent increase in eosinophil count eosinophil count > 1500 cells/mm3
- CHF (dyspnea)
  - Restrictive Cardiomyopathy
  - Mitral regurgitation
- Systemic embolization

Eosinophilic Heart Disease
4 Stages:

1) Acute inflammatory myocarditis
2) Eosinophil rich thrombus deposition
   - Mediated by injured endothelium
3) Endocardial thickening
   - Valve involvement
4) Fibrosis



Adapted from Hirota Y: In Abelmann WH, Braunwald E [eds]: Atlas of Heart Diseases. Vol 2. 1995

Hypereosinophilic Syndrome (HES)

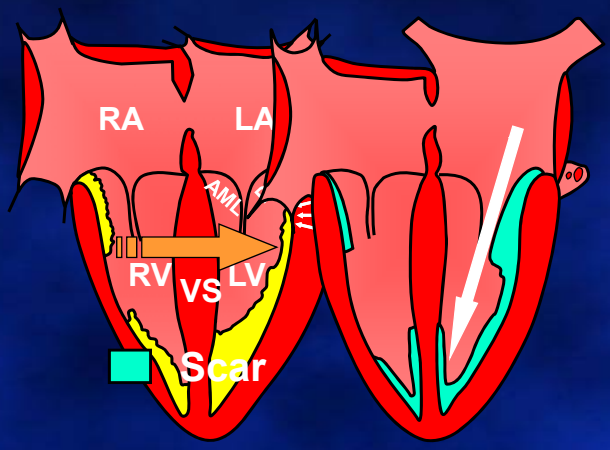
Cardiac Involvement: 40-60% of patients

2-D Echo & Doppler Findings

- LV > RV inflow apical thrombo-obliteration, endocardial thickening
- Restrictive diastolic dysfunction
- Subvalvular thrombosis, leaflet entrapment MV > TV Leaflets; MR&TR

Ommen, Am J Cardiol 2000

Natural History Hypereosinophilic Syndrome

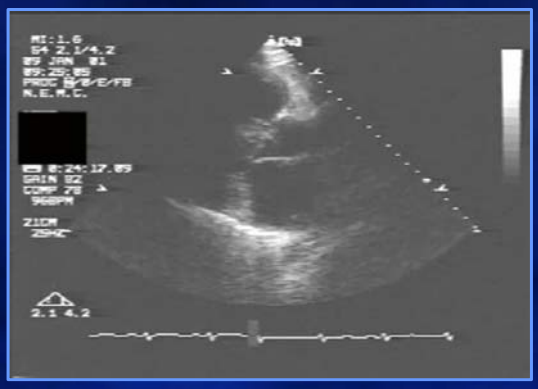


Myocarditis → Thrombus → Fibrosis

MAYO CLINIC

Image courtesy of Leslie Elvert RDCS

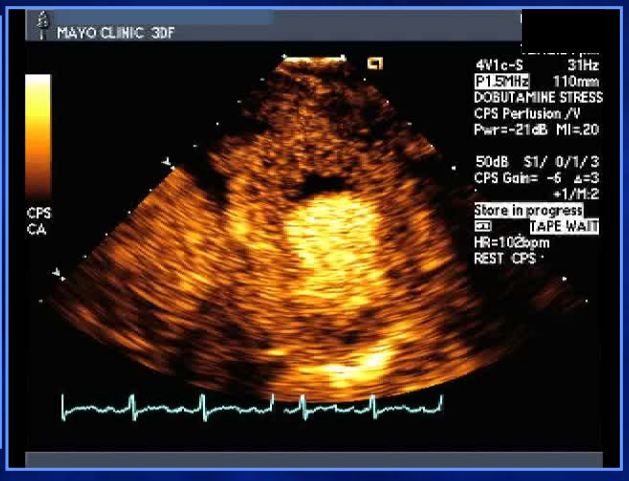
Basal LV Fibrosis with Mitral Posterior Leaflet Tethering



- Courtesy of Dr. Natesa Pandian

MAYO CLINIC

Eosinophilic Heart Disease
Contrast Helpful



Hypereosinophilic Syndrome

Treatment

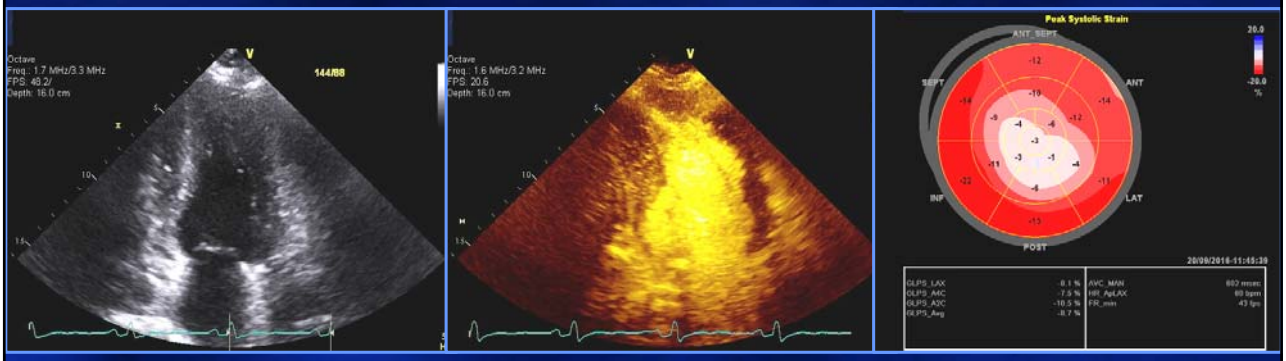
- Medical therapy
  - Corticosteroids
  - Hydroxyurea
  - Interferon
  - CHF Meds
- Surgical Therapy
  - Palliative

Echo Differential Diagnosis

- Apical hypertrophic CM
- LV Noncompaction
- LV tumor
  - Myxoma
  - Papillary fibroelastoma
- Ischemic LV dysfunction with apical thrombus

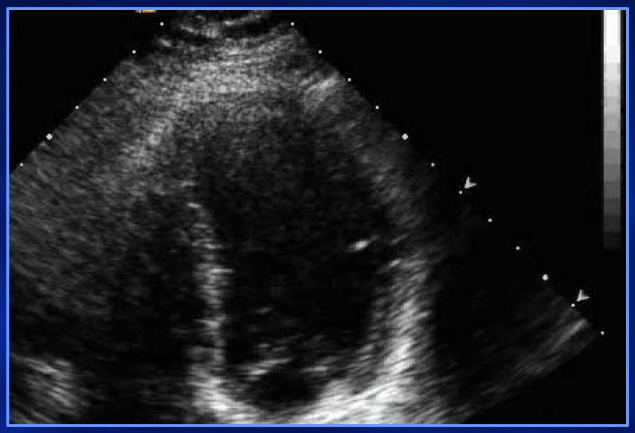
Differential Diagnosis: LV Myxoma

Differential Diagnosis: Apical HCM

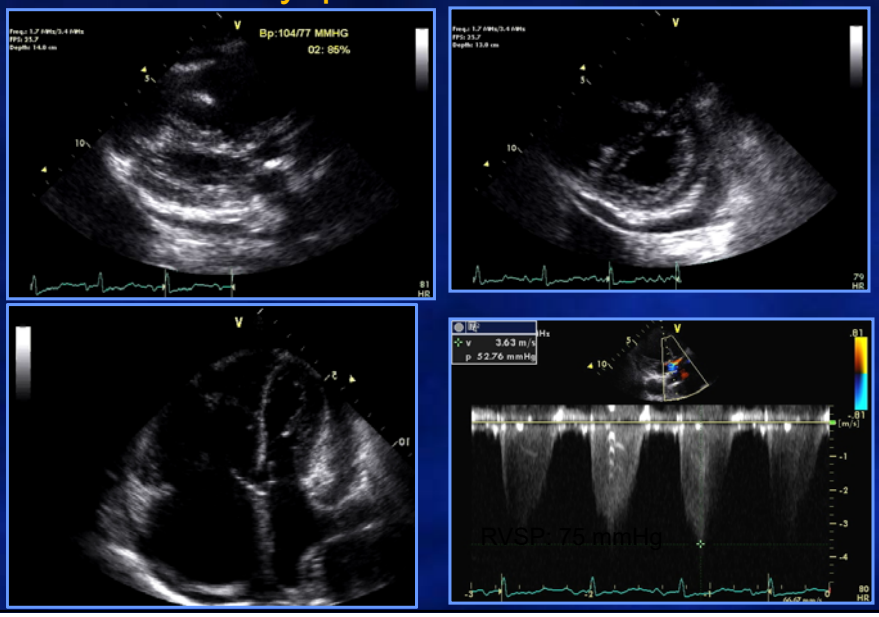


Our Case:

TTE after 2 months of anticoagulation
and 1 month of prednisone therapy



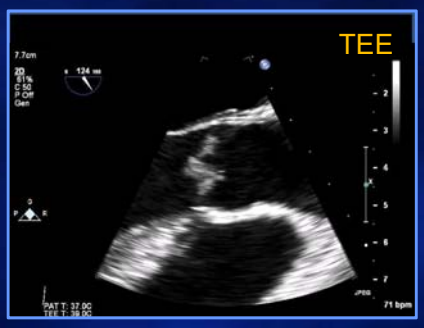
Patient with CREST Syndrome:
Dyspnea and Edema



Scleroderma and Pulmonary HTN

- PH present in 8-12% of scleroderma patients
  - Higher risk in CREST patients
- Accounts for 30% of deaths
- Screening for PH recommended
- RV dysfunction, cardiac index and pericardial effusion are markers of poor prognosis in PH

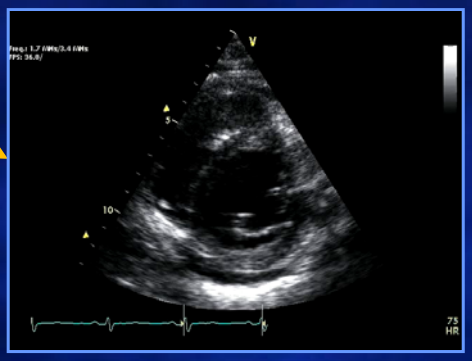
33 Year Old Female → Multiple Strokes



- ANA positive and Antiphospholipid antibodies present
- Libman-Sacks endocarditis

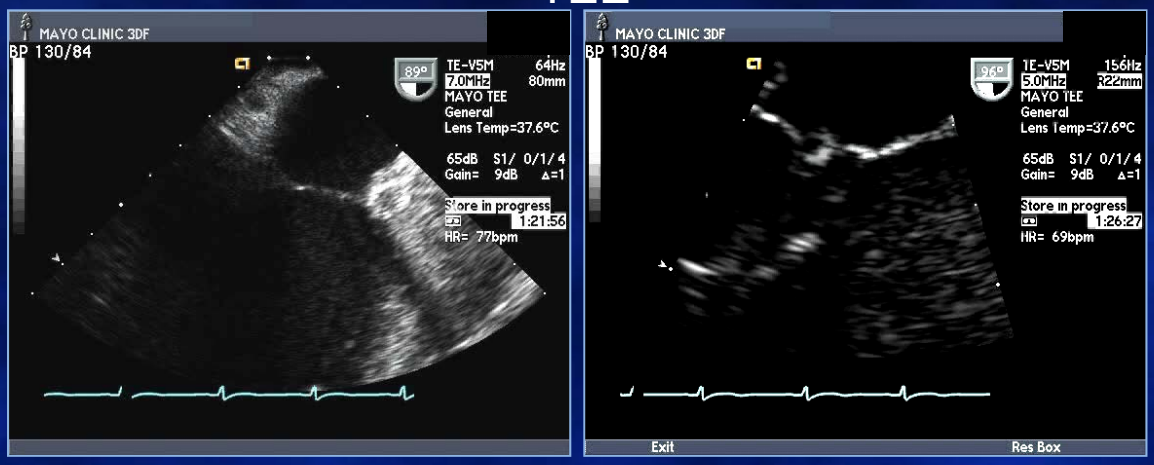
Systemic Lupus Erythematosus Cardiac Involvement

- Pericarditis (fluid ANA+)
  - 50-60% of cases have pericardial effusions
- Lupus anticoagulant
- Anticardiolipin or Antiphospholipid Abs
- Myocarditis
- Coronary arteritis
- Libman-Sacks (Marantic) vegetations



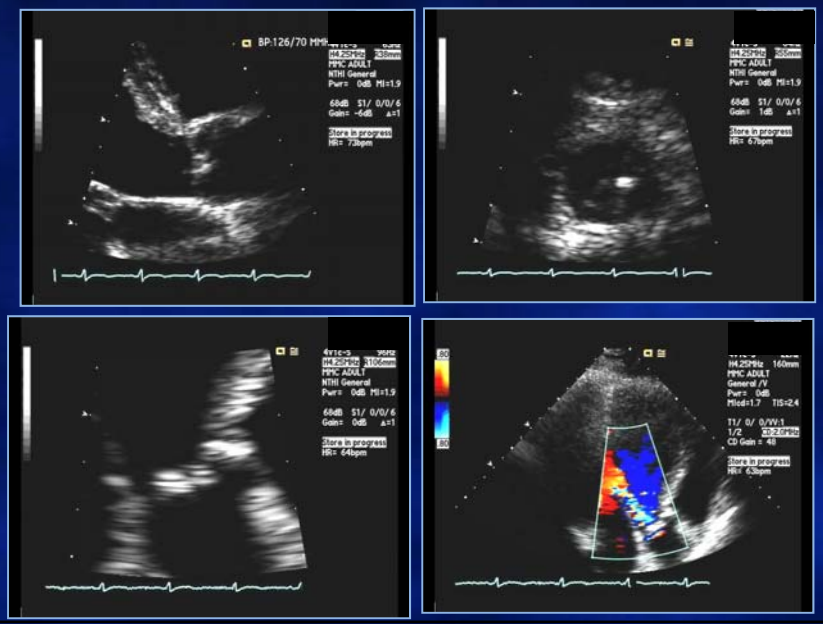
18 y.o. Female with Occipital Stroke

TEE



- Lupus anticoagulant + antiphospholipid antibodies present
- Libman-Sacks endocarditis

Not only the mitral valve!



Antiphospholipid Syndrome
Diagnosis confirmed at surgery

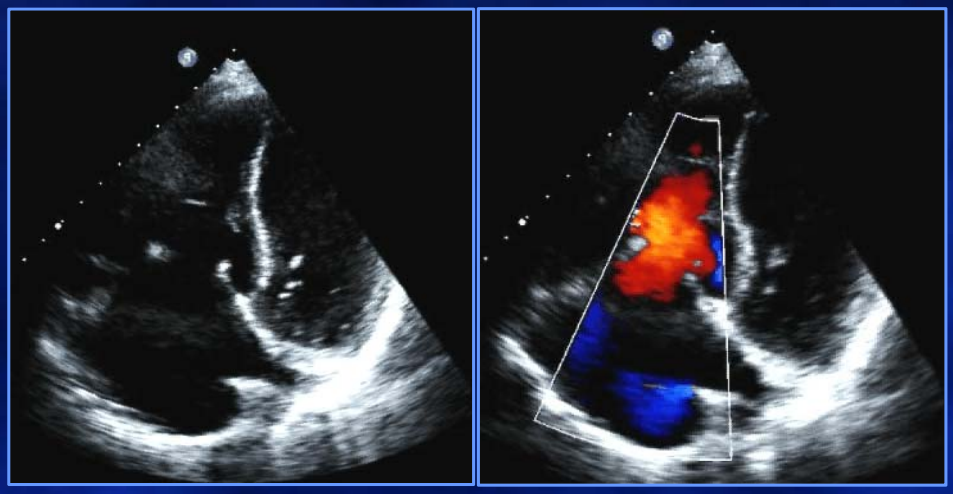
- IgG and IgM Antiphsopholipid antibody
- Importance of recognition
  - Unlikely repair
  - Choice of prosthesis??
  - Anticoagulation??

Systemic Lupus Erythematosus
Cardiac Involvement

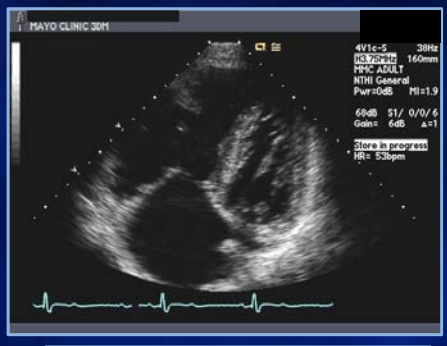
- Pericarditis (fluid ANA+)
- Lupus anticoagulant
- Anticardiolipin antibodies
- Myocarditis
- Coronary arteritis
- Libman-Sacks (Marantic) vegetations

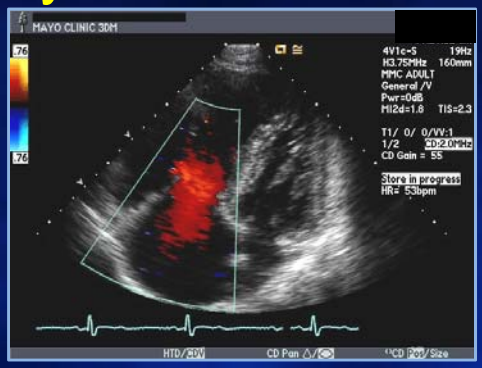
Courtesy of W Edwards MD

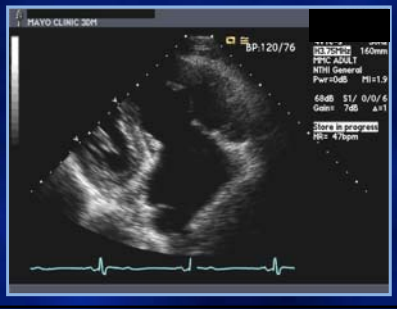
39 year old male with diarrhea, flushing and weight loss

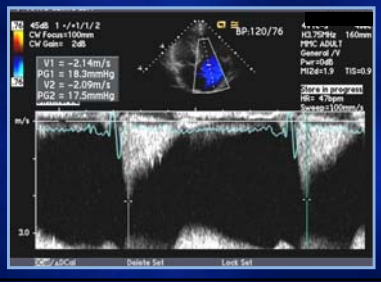


Carcinoid Syndrome

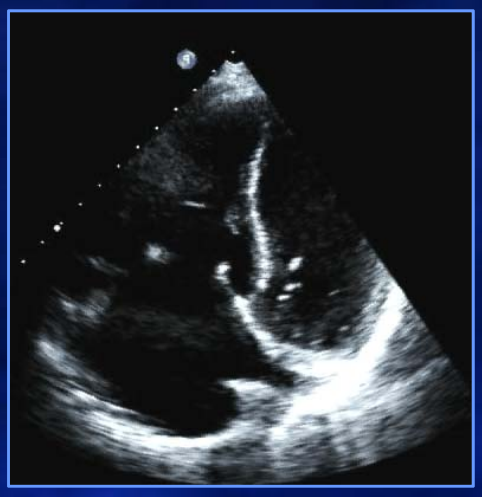








Carcinoid: Echo Features



Tricuspid valve

- Thickened leaflets
- Retracted leaflets
- Fixed semi-open position

Pulmonary valve

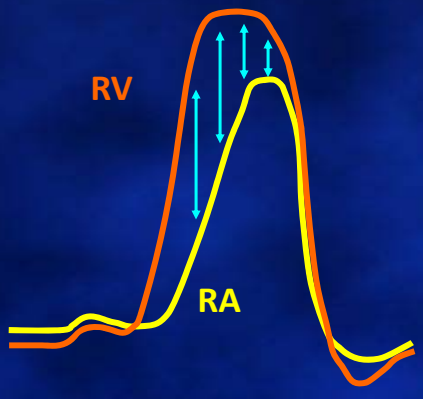
- Thickened cusps
- Retracted and rigid

Connolly HM. Curr Cardiol Rep. 2006

Severe (Torrential) Tricuspid Regurgitation

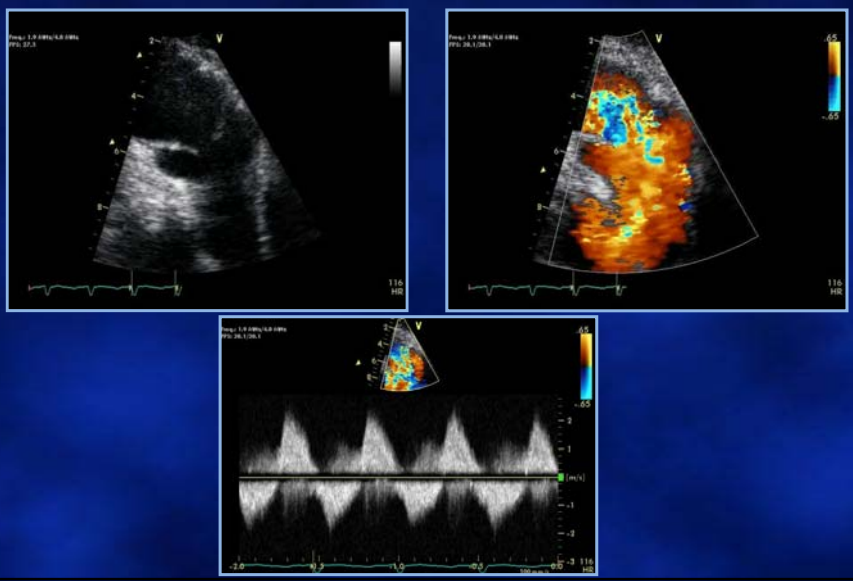
Systolic RV → RA pressure equalization

TR CW Doppler

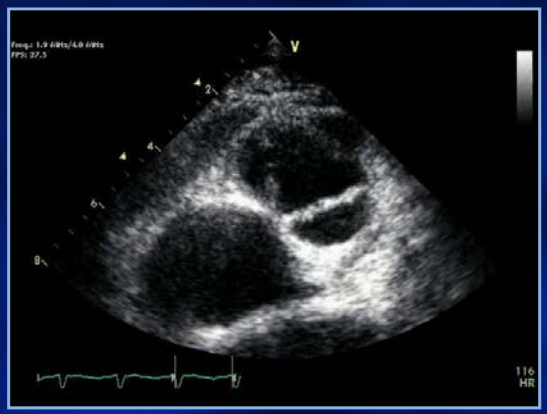


Courtesy of Dr. WK Freeman

Pulmonary Valve Involvement



Pulmonary Valve Involvement



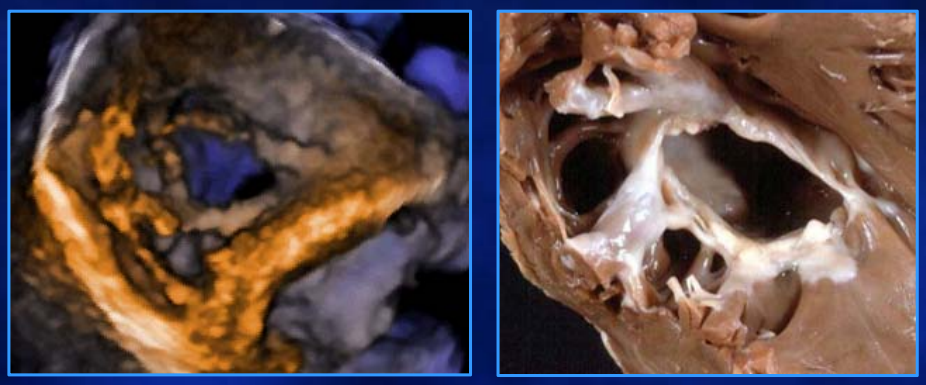
Adapted from Mayo Image Data Base, William Edwards, MD

Carcinoid Heart Disease

- Carcinoid tumors: 1-2/100,000
- Carcinoid syndrome in 20-30%
- Deposition of a matrix-like material on the valves and endocardium of the right side of the heart
- Treatment of tumor does not cause regression of valve disease

Connolly HM. Curr Cardiol Rep. 2006

Carcinoid Syndrome: 3D TTE



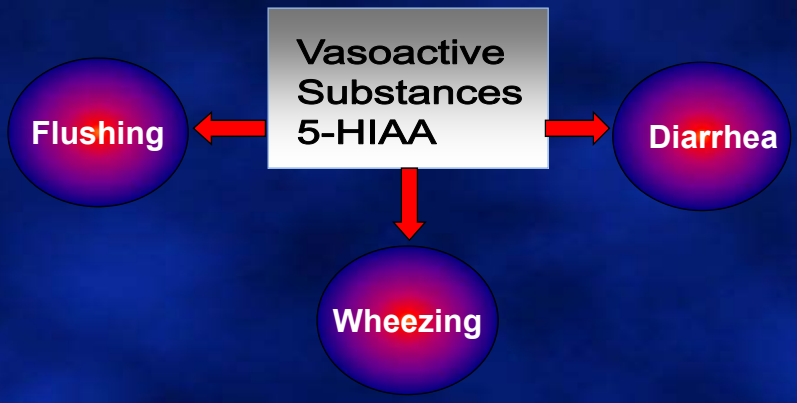
Courtesy of Denisa Muraru, MD, PhD
Padua, Italy

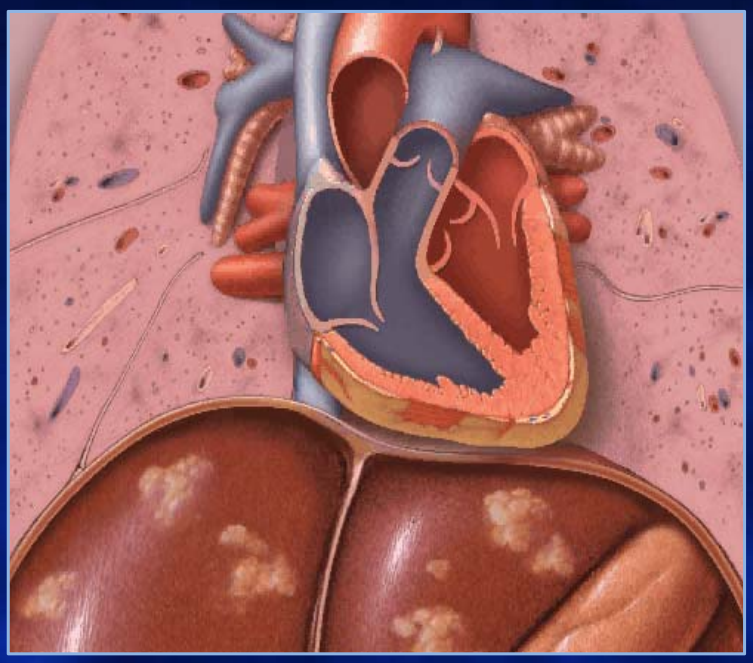
Eur Heart J Cardiovasc Imaging 2012

Carcinoid Heart Disease

Over 50% of patients with Carcinoid

Syndrome develop cardiac involvement





Courtesy of Dr. Heidi Connolly

410

JACC Vol. 25, No. 2
February 1995:410–6

Outcome of Cardiac Surgery for Carcinoid Heart Disease

HEIDI M. CONNOLLY, MD, FACC, RICK A. NISHIMURA, MD, FACC, HUGH C SMITH, MD, FACC, PATRICIA A. PELLIKKA, MD, FACC, CHARLES J. MULLANY, MD LARRY K. KVOLS, MD

Rochester, Minnesota

Objectives. The hypothesis was that cardiac surgery for symptomatic carcinoid heart disease in conjunction with adjunctive therapy could improve the long-term outlook of patients with carcinoid heart disease.

Background. Patients with carcinoid heart disease have a dismal prognosis; most die of progressive right heart failure within 1 year after onset of symptoms. Improved therapies for the systemic manifestations of the carcinoid syndrome have resulted in symptomatic improvement and prolonged survival in patients without heart disease.

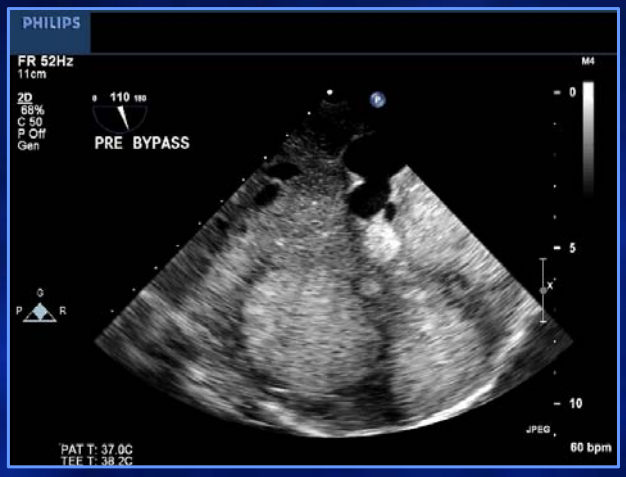
Methods. Twenty-six patients with symptomatic carcinoid heart disease underwent valvular surgery. Preoperative clinical, laboratory, Doppler echocardiographic and hemodynamic factors were evaluated. The survival of the surgical group was compared with that of a control group of 40 medically treated patients.

Results. There were nine perioperative deaths (35%), primarily from postoperative bleeding and right ventricular failure. Of the 17 surgical survivors, 8 were alive at a mean of 28 months of follow-up. The postoperative functional class of the eight surviving patients was substantially improved. Late deaths were primarily due to hepatic dysfunction caused by metastatic disease. The only predictor of operative mortality ($p = 0.03$) was low voltage on preoperative electrocardiography (limb lead voltage ≤5 mm). Predictors of late survival included a lower preoperative somatostatin requirement and a lower preoperative urinary 5-hydroxyindoleacetic acid level. There was a trend toward increased survival for the surgical group compared with the control group.

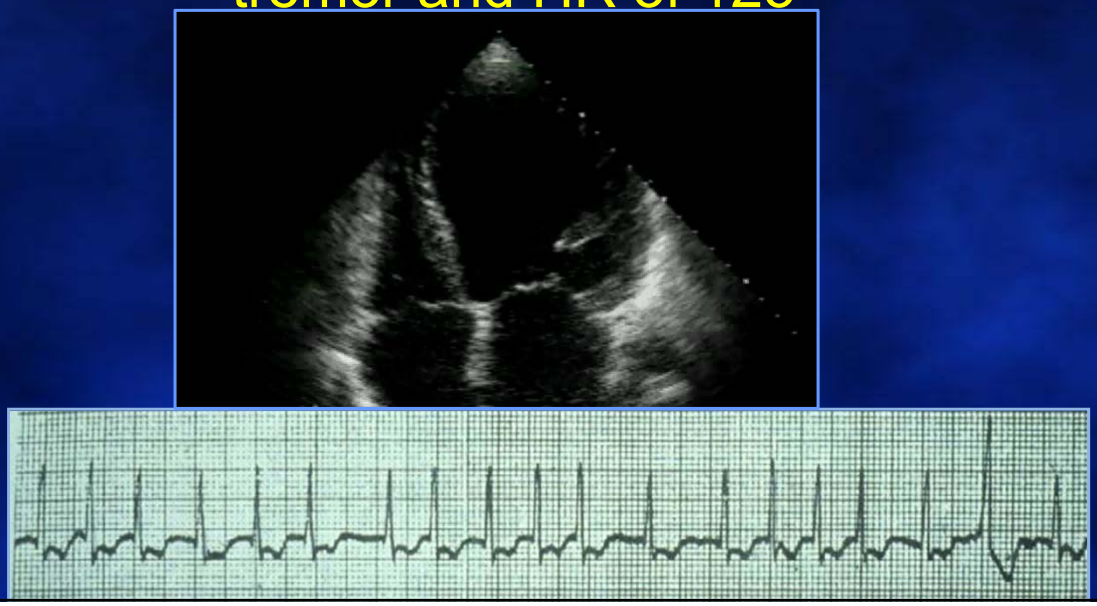
Conclusions. Because new therapies have improved survival in patients with the malignant carcinoid syndrome, cardiac involvement has become a major cause of morbidity and mortality. Valve surgery is the only definitive treatment. Although cardiac surgery carries a high perioperative mortality, marked symptomatic improvement occurs in survivors. Surgical intervention should therefore be considered when cardiac symptoms become severe.

(J Am Coll Cardiol 1995;25:410–6)

Carcinoid Tumor: : Liver Metastases



58 yo woman with weight loss, tremor and HR of 125



Hyperthyroidism

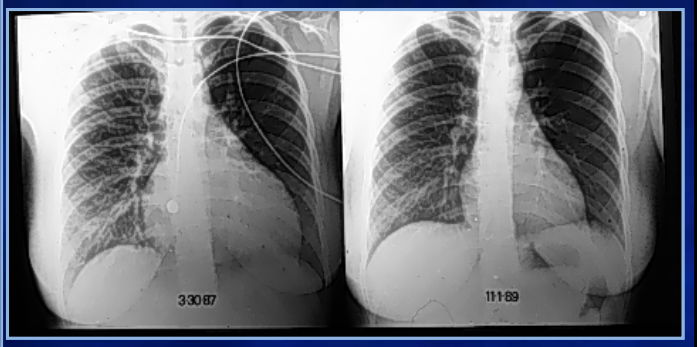
- Atrial fibrillation
  - difficult to rate control
- Decreased Peripheral resistance
  - hypotension
- Exacerbation of underlying CAD
  - increased myocardial O2 demand
- Tachycardia induced cardiomyopathy

Tachycardia Mediated Cardiomyopathy

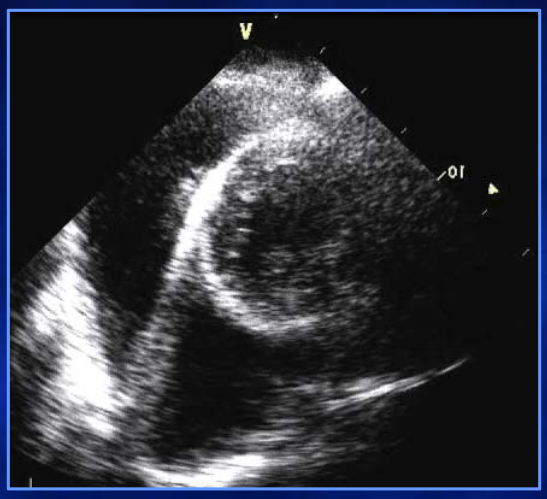
- 25% of patients w/ LV dysfunction & AF will have improved EF with rate control
- Usually *unaware* of rhythm
- Resting heart rate - poor indicator of overall rate control
- Consider in all pts with AF & LV dysfunction

Grogan M et al. AJC, 1992.

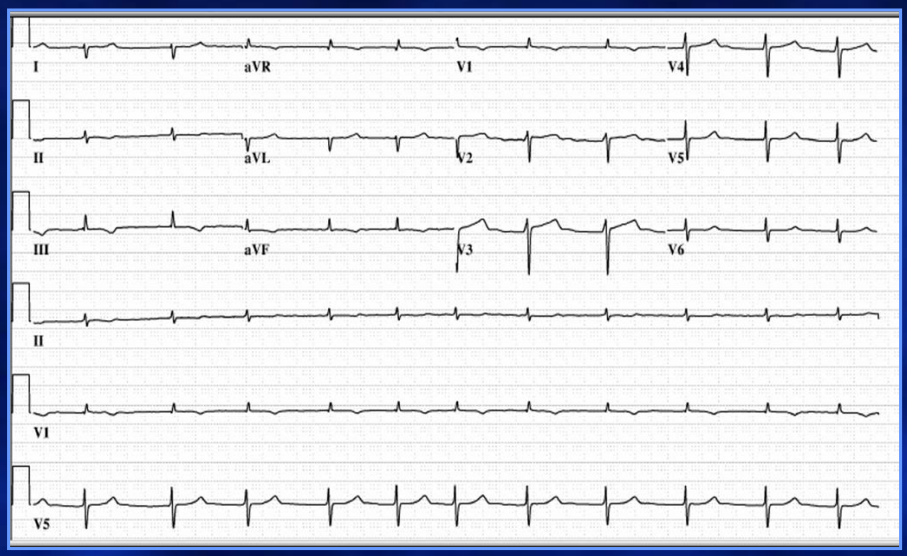
2 Years after Cardioversion and Treatment of Hyperthyroidsin



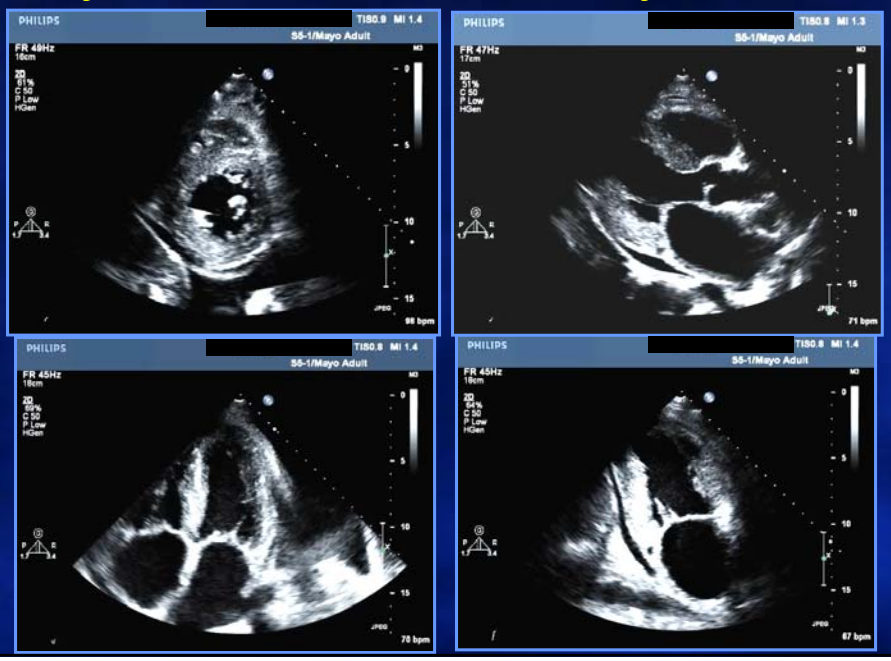
Hypothyroidism:
Large Pericardial Effusion



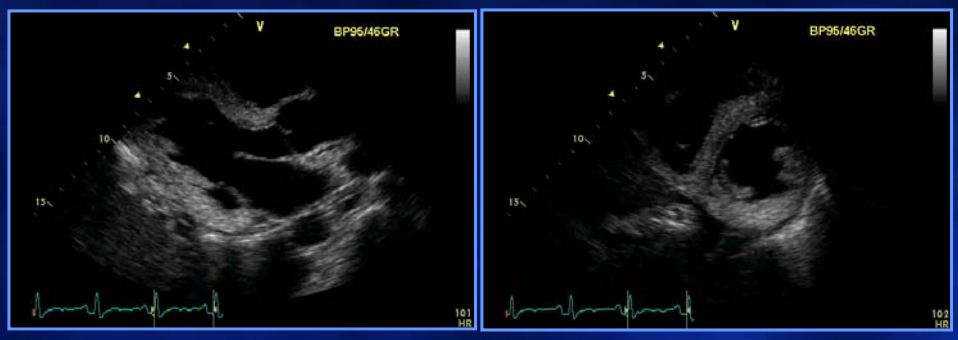
43 year old man



43 year old man with amyloidosis

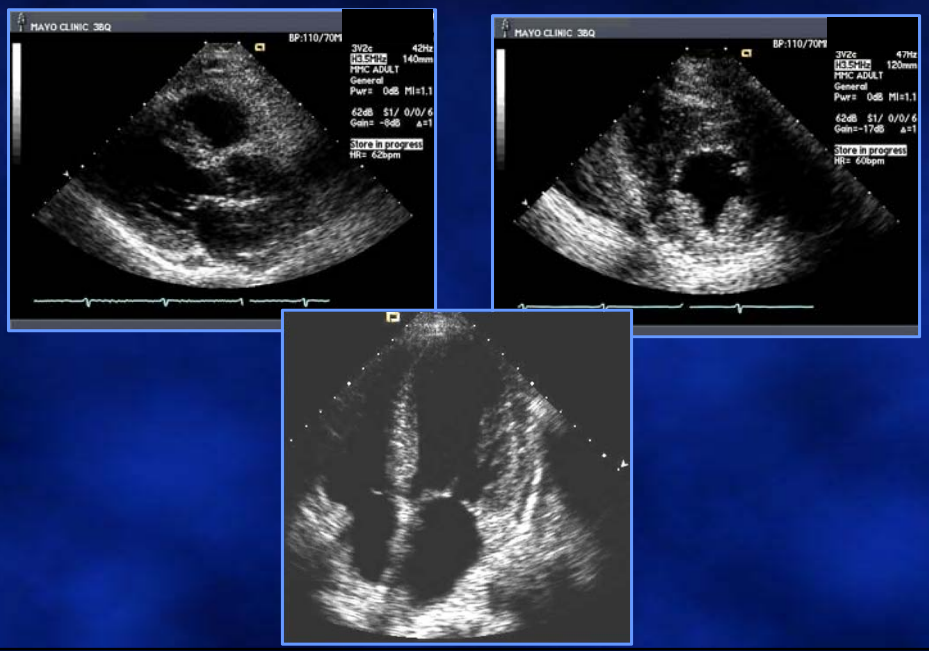


Primary Hyperoxaluria



- Rare metabolic disorder with autosomal recessive inheritance
- PHO type 1 (0.11 - 0.26 per 100,000 live births)
- Enzymatic defect resulting in enhanced conversion of glyoxalate to poorly soluble oxalate which is excreted in the urine

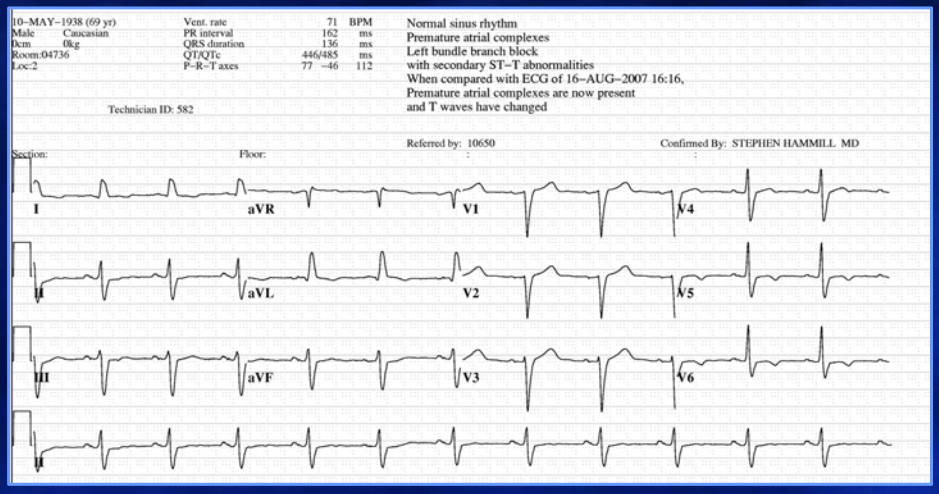
Hydroxychloroquine-induced Cardiotoxicity



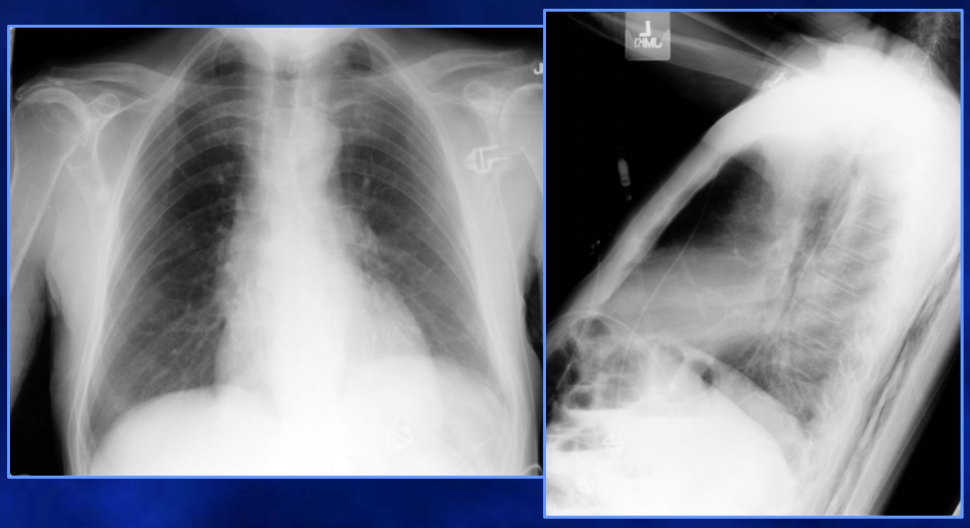
A 60 year old male farmer is referred for evaluation of dyspnea

- NYHA Class III symptoms
- PMH: Type 2 DM
- Abnormal LFT's
- Physical Exam:
  - 110/70 mmHg, HR 70 BPM
  - S3 gallop
  - Bronze skin

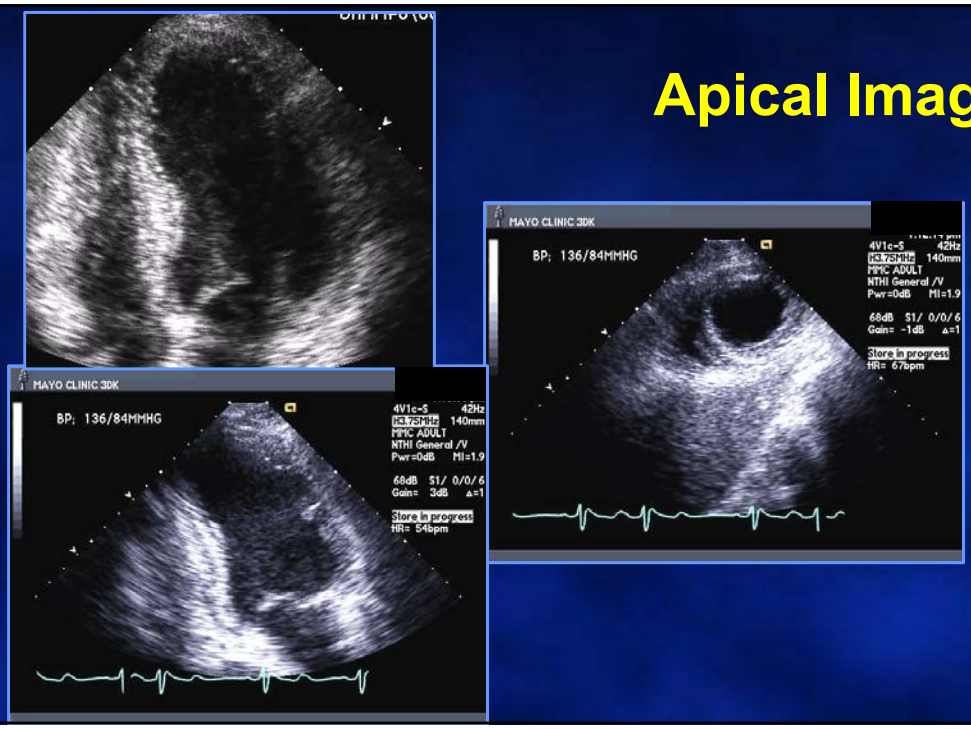
EKG



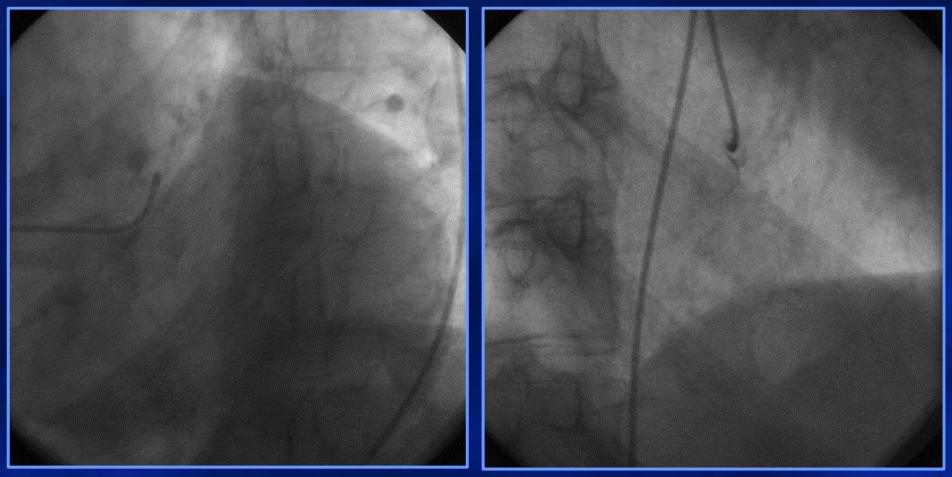
CXR



Apical Images



Coronary Angiography

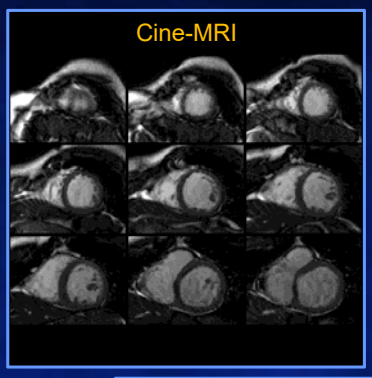


60 year old male farmer with Type 2 DM, bronze skin, and abnormal LFT's

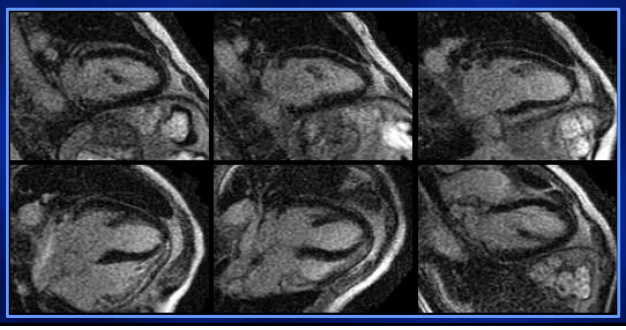
What is the most likely diagnosis?

a. Cardiac hemochromatosis
b. Cardiac amyloidosis
c. Cardiac sarcoidosis
d. Fabry's Disease
e. Carcinoid syndrome

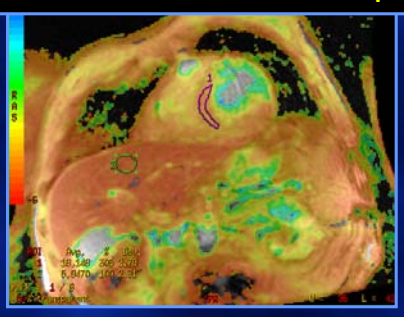
Cardiac MRI



No Delayed hyperenhancement



- The evaluation of the T2* relaxation time is an excellent noninvasive correlate of myocardial iron deposition and is a useful technique to follow response to iron-chelation therapy.
- Myocardial T2* has been shown to have no relation to serum ferritin and liver iron overload.
- T2* relaxation time predicts CHF and Arrhythmias



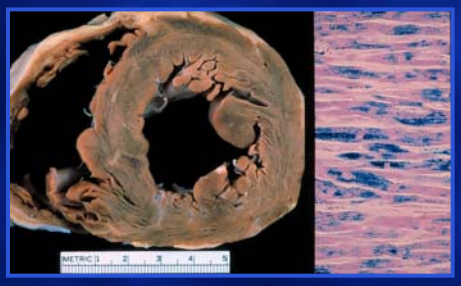
- This patient had a T2* relaxation time of 20ms that suggests hemochromatosis

Circulation 2009;120:1961-8
Eur Heart J 2001;22:2171-9.

Hemochromatosis

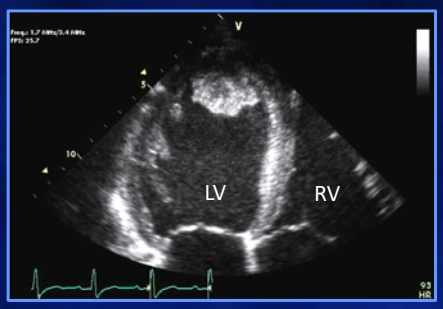
- ↑ total body iron – intracellular deposits in heart, liver, pituitary, pancreas, gonads, skin
- Think of this when DCM seen in setting of hepatic dysfunction; diabetes, tanned skin
- Diagnosis is critical, since reversible
  - Males 9:1
  - 2-3/1000 population
  - Ferritin usually > 500, transferrin > 50%
- Normal wall thickness
- Arrhythmias, conduction abnormalities

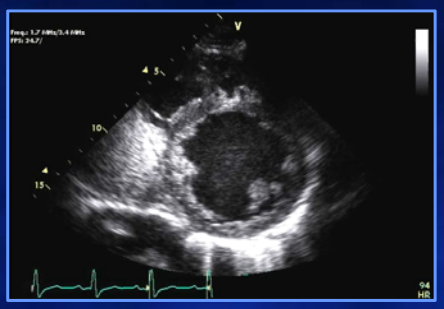
Intracellular iron
– directly toxic to myocytes

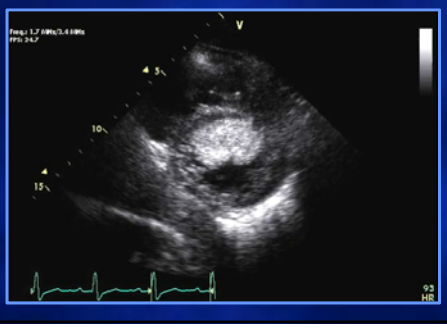


Courtesy of William Edwards, MD

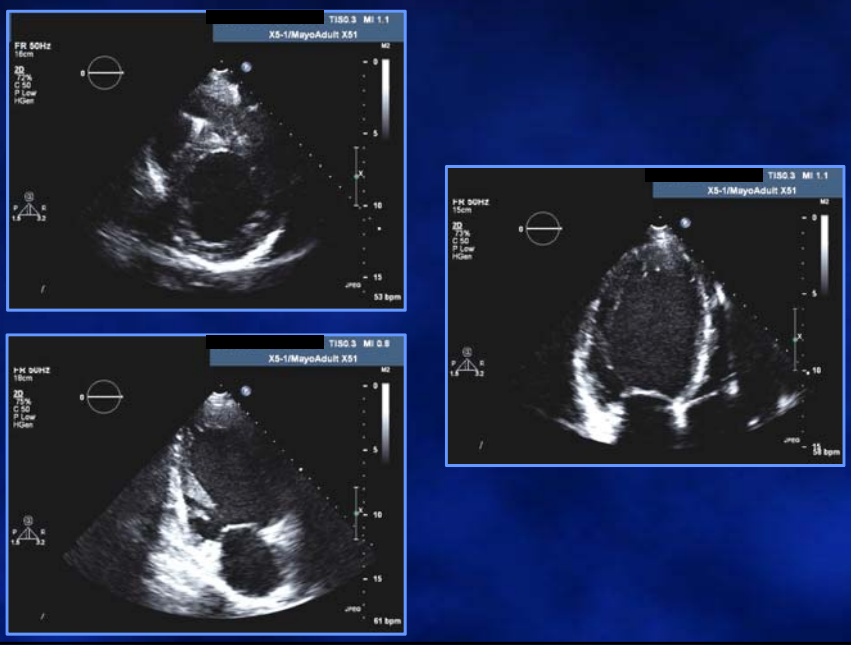
26 year old with Hemochromatosis







After Tx with Deferoxamine



Heomochromatosis: Take Home Points

- The Iron Heart is a weak heart…
- Hemochromatosis may be a cause of idiopathic dilated cardiomyopathy
  - Reversible with treatment
- Cardiac MRI (T2 relaxation time) is important in helping to establish diagnosis and monitoring treatment effects

Conclusions:

Systemic Diseases and the Echo Boards

- Carcinoid Syndrome
- Hypereosinophilic endomyocardial disease
- Sarcoidosis
- Systemic Lupus Erythematosus
- Scleroderma/Crest: Pulm Hypertension
- Amyloidosis
- Hyper or Hypothyroidism
- Radiation Heart Disease
- Drug Induced Valve Disease
- Hemochromatosis

Thank You!

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